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Reade - revalidation/rheumatology

Dr. S. A. de Waal, primary physician
(address)

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Reference number: -XXXXXX
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Cc: - BovenIJ Hospital, attn. Mr. A.L.M. Bakx, cardiologist
- BovenIJ Hospital, attn. Mr. M.M. Meijs, dermatologist

Dear colleague,

We saw your patient named above with urgency, at the request of colleague Bakx concerning Raynaud phenomenon.

Current medication: Fluticason nose-spray and oral contraception, since two days Naproxen.

History: one month ago a first attack of blue hands and blue discolorations in the neck, this disappeared again after a shower. Two weeks later a second attack of blue hands, the blue discoloration was as well present in neck and shoulders, chest, back and knees and elbows. This was not specifically due to cold. The blue spot around the neck is there now for two weeks, the blue discolorations on the hands disappeared again, and further did all else go away as well actually.
Since one month complaints of dyspnea when walking, no cough, no tickling *c*ough, can climb stairs but is panting then. Since one to two months progressively dizzy in attacks of max. 5 minutes. Since one week consistent headache, preventing sleep, frontal.
Two to three months ago *gecollabeerd* , that was finished this last month. Earlier a period of stomachache, over a few years, with diarrhea two to three times a week, but then twice a day thin as water but without blood or mucus, in addition nausea and stomachache. No tingling except in the arm yesterday. No other skin conditions like red spots, not psoriasis, not in family either. No alopecia, no sun-allergy other than during a vacation in Turkey.

Oral aphtae*,* but fewer recently and never vaginal aphtae. No risk of STD, recent tests for chlamydia were negative, no complaints of dryness in the eyes, or mouth. No inflammation of the eyes, no tick-bites other than very long ago. No crusts in the nose,

but a cold often, no ear infection. No fever, weight stable, tired for years. No obvious night sweats, but warmer than usual.
Review of systems significant for pain in the chest for the past three weeks, specifically shooting pains under left breast. Patient used to smoke a pack a day, recently this has diminished to a quarter to a half pack a day. Has about one drink per month, is in senior year of high school and lives at home. Her mother has fibromyalgia.

Physical examination:
Vital young woman, appears stated age with normal slender posture, one pale spot around the clavicle in the throat. BP110/65 mmHg, pulse 80/min. head and throat normal with, sputum present in mouth. No pathologic lymphadenopathy, normal heart tones, no wheezes or crackles over lungs, vesicular breath-sound in all fields, no crepitations. Abdomen normal peristalsis; liver and spleen not enlarged and not palpable, abdomen soft. On skin no signs of vasculitis, but a pale blue coloration in the throat, nowhere else. Arteria temporalis on both sides normally palpable, arteria radialis on both sides normally palpable, feet symmetrical hard to feel. Cold hands and feet, delayed capillair refill in the toes. No palpable purpura, no nail fold lesions.

Neurological examination:
Foot sole reflex symmetrical normal, Achilles tendon reflex and knee tendon reflex as well as biceps tendon symmetrical, not easy to bring forth, proprioception in the MTP joints intact on both sides.

Rheumatological examination:
No arthritis, somewhat sagging MTPs in further a bit of hyperlaxity. No particularities in the spine.

Laboratory examination of 12/9/2011 in BovenIJ Hospital: CRP 1 mg/l, normal values for leucocytes, and differential, thrombocytes, PTT and apt, dimer, creatine, liver enzymes.
Laboratory examination of 12/22/2011: BSE 3 mm/hr, normal values for hemoglobin, MCV, total eosinophils and leuco differentation, creatine, calcium, albumin, liver enzymes, LDH, CRP, complement C3 and C4, IgA, IgG and IgM, TSH, IgM RF and anti-CCP, ANA negative (cytoplasmic positive), anti cardiolipin antibodies, IgM and IgG negative, anti Beta 2 glycoprotein 1 negative, ANCA immune fluorescence negative as well as MPO and PR3 and Elisa negative, lupus anti-coagulant negative.

Urinalysis:
Other than a trace of ketones, no particularities, specifically no protein and no erythrocytes. Repeated urine examination on Jan 4th: leucocytes 0-5, bacteriae weak positive, again no proteins and no erythrocytes.

Conclusion and discussion:
Colleagues Meijs and Bakx found Raynaud phenomenon in the patient, not present in my examination, but there's still a cyanotic coloration around the clavicle. Despite above

described extensive diagnostics as well as a biopsy via dermatology where a little inflammation-remains was visible, no lymphocytic vasculitis and no hematoma and an ECG which was good.
With that at this point no clear diagnosis. The are no leads for palpable purpura - with that the diagnosis of cryoglobulinemia and hyperviscosity, anamnestic neither leads for cold injury of pressure related disorders, at this point no connective tissue disease, for thoracic outlet are the *livide* lesions too widely present.
Differential diagnostic then remains carcinoid, pheochromocytoma and Takayasu. But by patient blood pressure is normal. In first instance the patient will take a cuticle capillaries microscopy - if nothing appears from that, I consider sending her to an internist to exclude a carcinoid. The skin disorders should rather be red to purple and last max. 30 minutes. They can become carcinoid however. Diarrhea is present, dyspnea can belong to it as well.

With professional respect,

Dr. F. Turkstra, rheumatologist

Skin Biopsy:

Clinical data:DD/Acrocyanosis? Pigment? Perniosis? Traumatic/hematoma.
Since two weeks blue-ish maculae in throat, hands, elbows and knees.

Macroscopy:
GW/*biopsy punch* diameter 0.2 cm, depth 0.3 cm. Cass. 1: t.i.

Microscopy:
Serial cut skin biopsy clothed by a wavy epidermis with at the surface a thin basket-weave epidermis. The epidermis is small, shows basically normal stratification of the cores and toward the surface *normal riping.* Superficial in the dermis multiple thin-walled capillaries clothed by a single layer of endothelium-cells. In the lumen some erythrocytes. There is no evident extravasation of erythrocytes. No fibrionoid changes in vein wall.
Round veins just a few lymphocytes. No lymphocytic vasculitis. Deeper in the skin biopsy in the dermis some hair-follicles and glands. Around a single hair follicle some lymphocytic infiltrate. Deeper in the biopsy some sweat glands with focal as well a little lymphocytic infiltrate. No increase in iron-pigment. No hematoma. In the additional PAS-coloration a thin basal membrane. No fungus found. No increase of mastocyte cells. The EvG coloration shows some fragmentation of elastin-fibers. Superficial in the dermis a few melanophages.
Coupes have been co-judged by colleague M.M.S. Mulder.

Conclusion:
Skin biopsy of throat: skin with a light perifollicular inflammation and inflammation-rest.

No specific histological image, specifically no clues for a lymphocytic vasculitis. No hematoma.

Pathologist: Dr. M.J. Flens